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Case Report

Uterine Preservation and Vaginal Reconstruction in a Patient with Congenital Vaginal Agenesis Presenting With Cyclic Menouria

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ABSTRACT Herein we report the case of a patient with primary amenorrhea and cyclic menouria. The patient was a 20-year-old woman with primary amenorrhea and inability to achieve sexual intercourse. Clinical examination revealed normally developed labia majora and minora, clitoris, and external urethral orifice, but no vaginal opening. A mature female pubic hair pattern was present, and axillary hair development was normal. Breasts were normally developed. Abdominopelvic magnetic resonance imaging demonstrated a remnant upper vagina and unicornuate uterus filled with fluid, and left-sided renal agenesis. Intraoperatively, a congenital vesicouterine fistulous tract was observed. The fistulous tract was completely resected. Vaginal reconstruction using a sigmoid colon pedicled flap was performed. The proximal part of the neovagina was connected to the remnant cervix, and a Foley catheter was left in the uterine cavity for 7 days to prevent obstruction. The patient has been menstruating regularly since the operation. Menouria might be an early sign of congenital vesicouterine fistula. Resection of the fistulous tract with uterine preservation might be considered in patients with vaginal agenesis. Journal of Minimally Invasive Gynecology (2011) 18, 682–685 © 2011 AAGL. All rights reserved.

Keywords: Menouria; Primary amenorrhea; Vesicouterine fistula

Congenital vesicouterine fistula is an uncommon anomaly, typically manifesting with urinary incontinence or recurrent urinary tract infections [1–6]. Urinary tract anomalies may commonly accompany genital system anomalies because of their common embryologic origin from the intermediate mesoderm of the nephrogenic ridges. Complete or almost complete arrest of development of 1 müllerian duct may lead to unicornuate uterus, and congenital vesicouterine fistula might arise from early duplication of the ureteral bud and its incorporation into the müllerian duct caudal to the urogenital sinus [7]. Increased pressure from vaginal agenesis has also been considered an important etiologic factor in the formation of congenital vesicouterine fistulas [8].

Herein, we report the clinical findings and management in a 20-year-old woman with primary amenorrhea and cyclic menouria who had a congenital vesicouterine fistula.

Case Report

A 20-year-old woman had primary amenorrhea and inability to practice sexual intercourse. Medical history included that the patient had been passing menstrual blood in urine cyclically since menarche at age 13 years. She had never menstruated through the vagina. She further denied any history of urinary tract infection, pelvic pain, urinary incontinence, surgery, or foreign-body insertion. Family history was not significant.

Clinical examination revealed normally developed labia majora and minora, clitoris, and external urethral orifice,

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but no vaginal opening. A mature female pubic hair pattern was present, and axillary hair development was normal. The breasts were normally developed. Abdominopelvic magnetic resonance imaging demonstrated a remnant upper vagina and unicornuate uterus filled with fluid, and left-sided renal agenesis (Fig. 1). Vaginal reconstruction using a sigmoid colon pedicled flap was considered [9]. The procedure was approved by the Institutional Review Board of Akdeniz University, and the patient signed informed consent.

After premedication with intravenously administered midazolam and cardiac monitoring, the procedure was begun with the patient under intratracheal general anesthesia and in the lithotomy position. A urinary catheter was inserted. A Pfannenstiel incision was made. The abdominal and pelvic cavities were inspected for possible anomalies or disorders. The distal segment of the sigmoid colon was located and suspended. Approximately 14 to 17 mm of the sigmoid segment was selected and marked based on the sigmoidal artery. A fenestration was made on the mesentery near the marked sigmoid segment in the proximal end. The mesentery was divided as far as the base of the vascular pedicle. Side branches of the pedicle were located and coagulated or ligated. A similar procedure was repeated at the distal end of the sigmoid segment. The proximal and distal ends of the sigmoid were cut, and the sigmoid flap was isolated based on its vascular pedicle. The proximal and distal ends of the remaining sigmoid segments were introduced and anastomosed to each other in 2 layers to maintain gastrointestinal continuity. The unicornuate uterus was then observed and examined, and its relation to neighboring tissues was checked. It was confirmed that there was a connection between the uterus and the urinary bladder. A cystoscopic procedure was attempted, and a vesicouterine fistula was observed (Fig. 2). The fistula was cannulated, and its track was dissected and excised from both ends. These ends were sutured in 2 layers. A retrourethral and retrovesical pouch was created extending from the blind vaginal introitus to the abdominal cavity, first in cut and then in blunt fashion, taking care to preserve the urethra, bladder, and rectum. The harvested sigmoid flap was transposed to the created vaginal pouch based on its vascular pedicle (Fig. 3). The peritoneal end of the sigmoid tube was exposed to the cervical opening of the uterus and attached to it using interrupted polyglactin 210 sutures (Vicryl; Ethicon, Inc, Somerville, NJ) in 2 layers, with 3-0 and 2-0 stitches. A 14F Foley catheter was placed in the uterine cavity to prevent obstruction. Flap perfusion was checked and confirmed. The distal opening of the flap was attached to the introitus using polyglactin 210 2-0 stitches. At the end of the procedure, a vagina approximately 15 cm long had been formed (Fig. 4). All incisions were subsequently closed. Prosthetic mold insertion was used one month after surgery for testing the length of the vagina.

The Foley catheter was removed from the uterine cavity at 7 days after the operation. The patient started menstruating regularly on postoperative day 14 and for the last 10 months. She denies any further postoperative urinary tract symptoms.

Fig. 1

Magnetic resonance image shows a unicornuate uterus and vaginal agenesis.

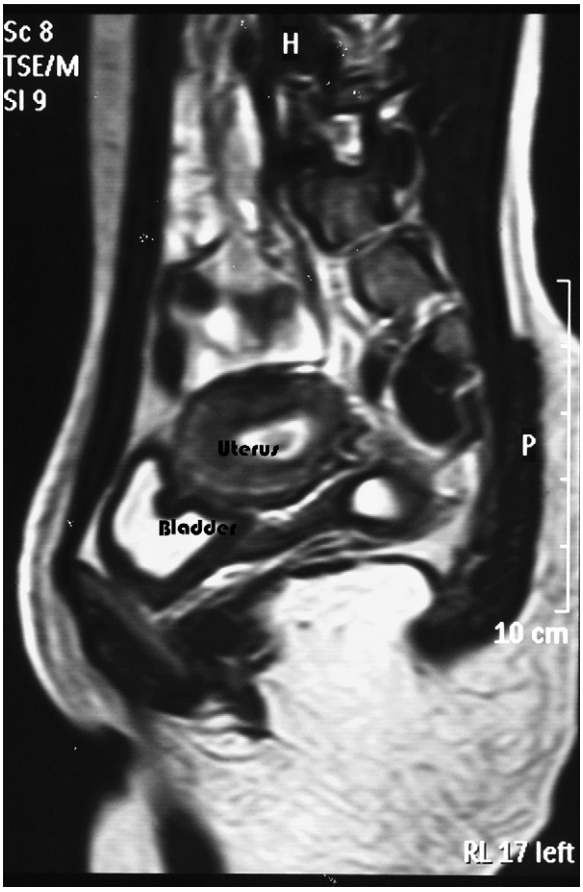

Fig. 2

Intraoperative appearance of the vesicouterine fistula tract.